Endoscopic Management of Esthesioneuroblastoma

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KEYWORDS

• Esthesioneuroblastoma • Olfactory neuroblastoma • Endoscopic
• Expanded endonasal approach • Skull base

KEY POINTS

- Esthesioneuroblastoma is a rare sinonasal malignancy presenting with nonspecific sinonasal complaints.
- Diagnosis is confirmed histopathologically, with characteristic small, round, blue cells in a neurofibrillary stroma with prominent microvascularity and lobular architecture.
- Higher histologic grade (Hyams) portends worse prognosis.
- Preoperative assessment and imaging are essential to guide surgical approach.
- Endoscopic endonasal resection is feasible in select cases with the goal of obtaining negative margins.

INTRODUCTION

Esthesioneuroblastoma (ENB), also known as olfactory neuroblastoma, is a rare malignant tumor of the nasal cavity first described by Berger and colleagues[1] in 1924. These tumors have a propensity for local invasion into surrounding structures and distant metastases, most commonly to the neck, lungs, and bones.

Patients with ENB typically present with nonspecific chief complaints of nasal obstruction and epistaxis, and definitive diagnosis is made on biopsy. Histopathology is consistent with lobular architecture, small round blue cells in characteristic Homer-Wright pseudorosettes and Flexner-Wintersteiner rosettes, and prominent microvascularity (**Fig. 1**). Hyams[2] developed a histopathologic grading system classifying ENB

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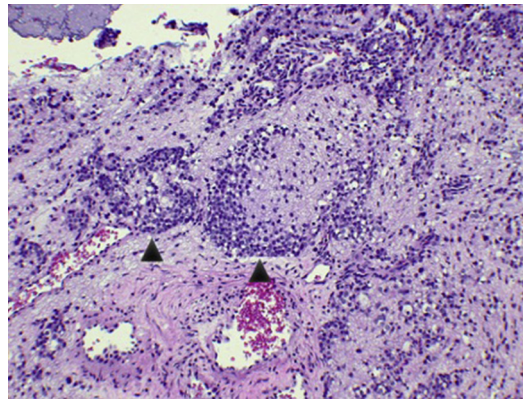

Fig. 1. Histopathologic sample of esthesioneuroblastoma. Note Homer-Wright rosettes (*arrowheads*) (H&E stain). (*Courtesy of* Dr James J. Sciubba, Lutherville-Timonium, MD.)

into four groups, with poorer prognosis occurring with increasing grade from I to IV (Table 1). Several attempts have been made to stage ENB based on imaging and surgical characteristics, with the most commonly used systems being developed by Kadish and coworkers,[3] Dulguerov and Calcaterra,[4] and Biller and coworkers,[5] with a more recent modification of the Kadish system by Morita and coworkers[6] (Table 2).

Management of ENB is generally surgical, with evidence suggesting that surgery with adjuvant radiation may provide the best prognosis.[7] Many surgical approaches have been described, including extracranial approaches,[8] craniofacial resection,[9,10] endoscopic-assisted craniofacial resection,[11,12] and most recently purely endoscopic expanded endonasal resection.[13,14] This article describes purely endoscopic surgical management of ENB and reviews outcomes of this approach described in the literature.

TREATMENT GOALS AND PLANNED OUTCOMES

Like other malignant sinonasal tumors, treatment goals and expected outcomes depend on extent of disease and tumor grade at presentation. Overall 5- and 10-year survival rates based on Surveillance, Epidemiology, and End Results tumor

Table 1
Hyams grading system

Microscopic Features	Grade I	Grade II	Grade III	Grade IV
Architecture	Lobular	Lobular	±Lobular	±Lobular
Pleomorphism	Absent/slight	Present	Prominent	Marked
Neurofibrillary matrix	Prominent	Present	May be present	Present
Rosettes	Homer-Wright	Homer-Wright	Flexner-Wintersteiner	Flexner-Wintersteiner
Mitoses	Absent	Present	Prominent	Marked
Necrosis	Absent	Absent	Present	Prominent
Glands	May be present	May be present	May be present	May be present
Calcification	Variable	Variable	Absent	Absent